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REMARKS ON JACKSONIAN EPILEPSY ILLUSTRATED BY AN UNUSUAL CASE.

BY JOHN SOUTTAR MCKENDRICK, M.B.

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THE case occurred in the wards of Sir William Tennant Gairdner, K.C.B., while I acted as his resident, and I have to thank Sir William for permission to publish the case, and for the interest he has taken in this paper.[1]

It is not my intention to give a critical description of Jacksonian epilepsy, on which so much has been written, but, in order to increase the interest of the case now reported, I shall discuss shortly some of the more important points in regard to the history, physiology, and pathology of this disease.

At the outset I may state that I have made free use of an admirable article by Rauzier which appeared in the January number of *La Semaine Médicale*, 1893, and which contains a valuable epitome of the literature of the subject up to that time (1).

Another review, likewise full of information, is written by Burlureaux in the *Dictionary of Medical Sciences.* Interest in the subject is also aroused by the perusal of individual cases described by such masters as Hughlings Jackson (2), Charcot (3), Ferrier (4), Dreschfeld (5), Ross (6), Gowers (7), Broadbent (8), and Byrom Bramwell (9). French writers have added largely to the literature of the subject, among whom may be mentioned Bravais (10), Fournier (11), Girard (12), Greffier (13), Pitres (14), Charcot et Pitres (15), and Bourneville (16).

[1] I have also to thank Dr. Ferguson for a pathological report, and Dr. Fred Dittmar for making careful sections of the brain tissue for me. To Dr. Samuel Prior I am indebted for the accompanying photomicrographs of the cerebral cortex, and to Dr. Hutton for his report on patient's condition on last admission.

History.

The medical history of convulsions dates back to the earliest times, and cases were described when only one side of the body, and indeed where only individual members were affected, but then *post-mortem* examinations, I suppose, were never made, and consequently no definite relation existed in the minds of writers between the central nervous system and the convulsive storms that occurred at various parts of the body. From the beginning of this century on to the middle of it, many cases were reported where convulsions affected certain parts of the body, and where definite organic lesions existed in the form of tumours, cysts, thickened membranes, &c., in the opposite side of the brain from the convulsed side, and yet these authors, both from want of experimental proof, and also from want of scientific reasoning, did not group the facts together in such a way as to establish a connection between the lesion and the convulsive seizures which were so accurately described.

Abercrombie (17), for example, describes as a class certain cases "distinguished by convulsions without any affection of the senses," where, he says, the convulsions may be confined to one side of the body, and he notes that in one case "there was a tumour on the surface of the opposite hemisphere, and in another a portion of the opposite hemisphere was indurated." He then, in fact, described a typical case of what we now call Jacksonian epilepsy, although he probably considered the presence of the tumour and induration as an accidental coincidence, and not the essential cause of the paroxysmal storm. In the same way Burlureaux (18) tells us that Parent-Duchatelet, Demongeret, Papavoine, Andral, and others, described cases of unilateral convulsions commencing in the fingers and corresponding with lesions of the ascending frontal convolutions of the opposite side, but they were entirely unaware of the pathological significance of these lesions.

The various forms of unilateral convulsions were first

discussed by Prichard in 1822, when he called them local convulsions. Bravais (19), in 1827, called the condition hemiplegic epilepsy, as he had frequently noted paralysis to be a sequel to the convulsion on the affected limb. Elliotson (20), in 1831, designated the disease partial epilepsy, where only a part of the body was convulsed, so as to distinguish it from idiopathic epilepsy, which was more general. This term is frequently used to-day.

In 1861, Hughlings Jackson discussed the disease in the most suggestive manner, and after careful investigation so clearly and accurately argued on the facts that he set on foot a train of investigation which culminated in the well-known researches of Fritsch, Hitzig, and Ferrier, on the localisation of the motor areas in the brain. From that date the disease has been happily called "Jacksonian Epilepsy."

This form of epilepsy has a still wider nomenclature, according as the symptoms differ somewhat from thôse usually described by Hughlings Jackson, and from the frequent association of certain pathological conditions which give rise to it.

Charcot called one form "a partial tonic epilepsy," where, dependent on an organic lesion in the motor area of the brain, a group of muscles were in a state of tonic contraction, rather than in convulsion. Another form he designated "vibratory epilepsy," where there was a constant shaking of the muscles affected during the tonic stage. A third variety he speaks of as "l'épilepsie partielle sensorielle," where a severe ophthalmic headache accompanies the convulsive seizure.

As syphilis is such a frequent cause of these convulsions it has been often called "syphilitic epilepsy." Numerous are the writers that have approached the subject from this standpoint, more notably Fournier (21), Charcot (22), Barbier (23), and Mallet (24). Lastly, it has been termed by some "apoplectiform epilepsy," because the patient, the subject of true Jacksonian epilepsy, had occasional apoplectiform seizures. True Jacksonian epilepsy and apoplectiform seizures have been known to alternate in some instances,

when the disease has received the name of "congestive apoplexy."

Physiology.

Hughlings Jackson, in his numerous writings, discussed the physiology and pathology of unilateral convulsions. Although at first he was not fortified by the results of physiological inquiry, he was of opinion that convulsions and hemiplegia were closely allied, and that the latter was merely the result of a more severe interference with the motor tract of the cerebral nervous system. Moreover, he concluded that convulsions were mere symptoms—the result of an over-excitability of certain motor nerve-cells, and although he was unaware of the particular nervous centres that were involved, apart from the fact that he had frequently seen *post-mortem* certain brain lesions on the opposite side of the brain, which he considered the cause of the irritation of the neighbouring brain substance, he was confident that different kinds of convulsions, as regards the part first affected, depended on irritative lesions affecting different parts of the brain cortex. In his own words, "a spasm beginning on one side implies local changes in the central nervous system as surely as one-sided palsy does;" and, again, "I [Hughlings Jackson] assert that there must of necessity be some place where the nervous system is diseased, or the spasm determined by causes acting generally would not be local."

He considered that there were centres for the face, arm, and leg, and that these were irritated in a definite order, just as he had noted that convulsions occurred in a definite order. He accordingly considered three varieties of convulsions:—

(1) Those beginning in the hand.

(2) Those beginning in the face and tongue.

(3) Those beginning in the foot.

It was not astonishing, then, that scientific observers were eager to submit these views to the test of experiment.

In Germany, Hitzig and Fritsch, and in England Ferrier, at once entered the field. Ferrier, in particular,

experimented on monkeys, and successfully localised certain motor areas in the cortex cerebri.

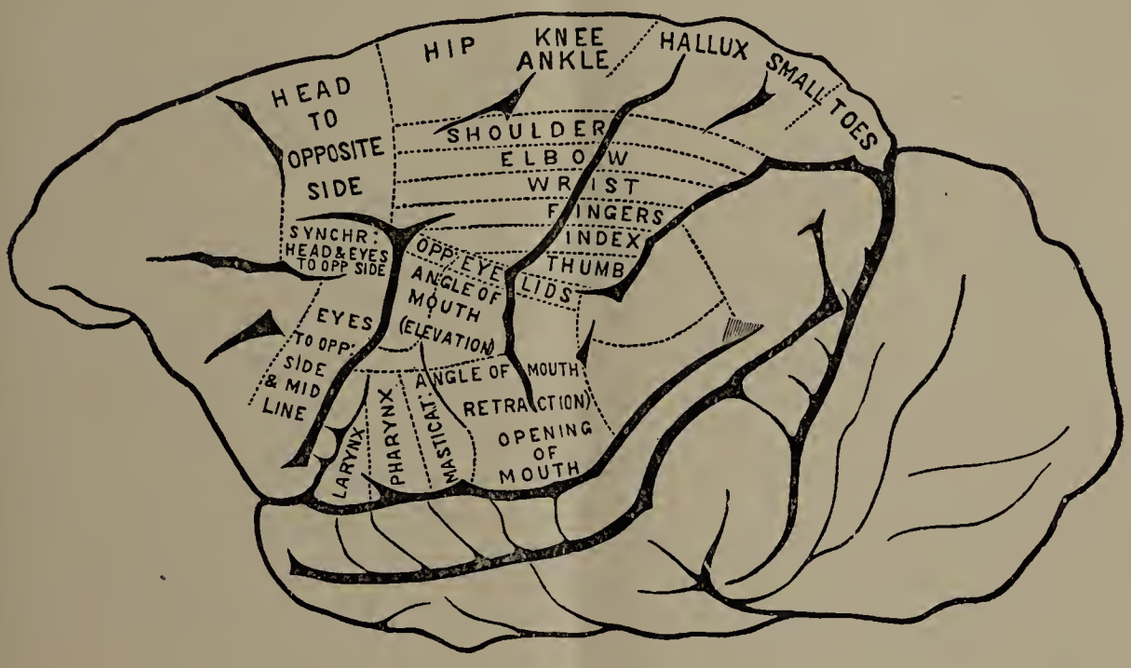
So indebted was he to Hughlings Jackson for the suggestion that led to this great research that he writes, "It is the great merit of Hughlings Jackson to have first clearly indicated the pathology of these affections" (convulsions) (25). His work on the "Functions of the Brain," published in 1876, Ferrier dedicated to Hughlings Jackson, "who from a clinical and pathological standpoint anticipated many of the more important results of recent experimental investigation into the functions of the cerebral hemispheres." A narrative by Ferrier, giving the experimental results, appears in the *Philosophical Transactions*, and at once there were two parties formed, the one side sceptical, the other full of belief.

Ferrier, however, was not the first to map out definite areas of the brain cortex that had to do with definite movements, for Fritsch and Hitzig (26) in 1870 discovered that the application of electrodes to an exposed part of the brain produced movements in certain parts of the body. There can be no doubt, however, that the results obtained by Ferrier were much more forcible and far-reaching than those of the German observers, and that they laid the basis of modern cerebral pathology, so far as motor areas are concerned.

The most notable of other writers who experimented in order to definitely localise particular cortical areas for definite movements were Beevor, Schäfer, Victor Horsley (27), Brown Sequard, Dupuy, and Burdon Sanderson.

For a time medical literature was teeming with the records of investigations by numerous experimentalists. When it was known definitely that particular areas in the cerebral cortex existed, which, when excited produced convulsions of particular muscles, and when destroyed caused paralysis without anæsthesia, clinicians and pathologists were ever on the look out for the discovery of brain lesions in the human subject that would correspond with definite symptoms observed during life. These efforts were richly rewarded.

We now know that the motor cortical area consists of a zone forming the convolutions around the fissure of Rolando, with pretty accurately defined areas for leg, body, arm, and face from above downwards. Irritation of these areas causes convulsions on the opposite side, and irritation of an area somewhat in advance of this causes deviation of the head and eyes to the opposite side (see diagram



—Beevor and Horsley). Having discussed briefly the physiology of unilateral convulsions, I intend now to make a few remarks on their pathology.

Pathology.

What is the pathological lesion, and how does this lesion cause irritation of the brain cortex? It has frequently been called the "discharging lesion."

The pathological lesions may be divided into two classes:

(1) Those that can be seen by the naked eye. (2) Those that can be seen only by the highest powers of the microscope. It is probable that lesions may exist that cannot at present be seen at all, or demonstrated by any known method.

Of the first class, any form of tumour, cyst, thickened membrane, a blood extravasation in or around the neigh-

bourhood of the fissure of Rolando, may cause convulsions, by irritation of the brain cortex.

This class practically includes all the forms of diseased brain tissue, meninges, and cranium in the region of the motor area of the brain. The various diseases have been enumerated by Rauzier (28), as follows:—

I. Affections of bone:—(*a*) tumours; (*b*) exostosis; (*c*) long splinter of bone.

II. Affections of meninges:—(*a*) tumours; (*b*) exudation, acute, subacute and chronic meningitis; (*c*) primitive hæmorrhages of meninges; (*d*) blood extravasations occurring in pachy-meningitis.

III. Affections of brain:—(*a*) Old and encapsulated abscesses; (*b*) cerebral sclerosis; (*c*) brain tumours; (*d*) hæmorrhagic foci; (*e*) ramollissement.

He lays special stress on traumatism and syphilis as causes in this form of unilateral convulsive seizure. An interesting table is appended by Rolland, who found that out of 112 cases forty-eight resulted from tumours, twenty-one from ramollissement, eight from traumatism, nine from superficial and deep aneurism, seven from cortical hæmorrhages, and five from cerebral atrophy.

The second class includes conditions in which there is possibly a change in the shape, size and number of the motor cells; or possibly from the help of modern histological methods of staining, a change may be detected in the protoplasmic substances of the brain cells; or the formation of a fatty substance around these cells; or possibly an interruption in the nerve fibres that proceed from these cells. There may be a minute change in the small arteries and veins that traverse the part—an arteriosclerosis. The origin or cause of these changed states of brain tissue is still obscure, but we must not overlook the fact that such minute changes in the motor area of the brain have been recognised with well-defined unilateral convulsions.

A greater puzzle in regard to the pathological significance occurs in certain cases where convulsions have been unilateral, and from a clinical standpoint entirely in

harmony with the convulsions described by Hughlings Jackson, and yet have presented no cerebral changes that could be discovered even by the highest powers of the microscope. These cases have been described as functional, but on physiological grounds we may hold that where there are well-defined unilateral convulsions there is in all probability a local lesion. Possibly, however, the disturbance may be of a chemical nature.

We yet know but little of the chemical constitution of a nerve cell, or of the chemical phenomena happening in it. One can conceive that an altered metabolism, and an altered nutrition or change in chemical state or chemical process, may produce marked change in function.

Those belonging to the first class (including tumours, &c.), then, produce convulsions of the opposite side of the body by irritating the surrounding nerve-substance by their presence. It may be by pressure on the brain substance direct, or on the small vessels: or it may be by causing an inflammation. The surrounding cells are in a state of tension, and when this is sufficiently high there is what may be called a molecular explosion. This view is borne out by Hughlings Jackson and Charcot; and, moreover, they consider that the paralysis resulting is in consequence of the weakened and exhausted state of these motor cells after the explosion has taken place.

It is difficult to argue out the pathology of those belonging to the second class (with only microscopical changes, &c.), unless we uphold a chemical theory. Hughlings Jackson has expounded a view bearing on this subject which I will quote: "Speaking in chemical language, the highly unstable grey matter of disease remains of the same constitution as the comparatively stable grey matter of health, but that it is of a different composition; and a further speculation is that the phosphorous ingredient is replaced by its congener nitrogen—that the nervous matter is more nitrogenised, and therefore more explosive" (29). In support of this view it may be pointed out that artificial explosives like gun cotton, nitro glycerine, and prussic acid, always contain a loosely combined molecule of NO_2. The presence of the nitrogen molecule implies instability.

The actual convulsion is frequently set agoing by some peripheral irritation, or disease of the abdominal or thoracic organs. It may be that certain poisons are circulating in the blood, which either pick out the motor area and cause convulsions, or more probably by producing a pathological change in the brain tissue or its vessels. It is easy to understand how these poisons would produce convulsions by acting reflexly, provided that an organic lesion is already present; but if no such lesion exists, it is difficult to conceive of a poison picking out the motor area more especially, and consequently producing convulsions.

Still, writers have considered that poisons, such as alcohol (30), lead (31), and urea (32) in uræmia are capable of selecting one motor area, and of producing typical unilateral convulsions. They have called these "toxic epilepsies."

With these remarks on the history, physiology, and pathology of Jacksonian epilepsy, I shall now proceed to a description of a case, which I take to be one of this type of disease, although in many points it is not typical. I shall endeavour after its description to enter into a short discussion on its more important and interesting features.

E. McG., aged 38, a housewife, was admitted into the wards of the Western Infirmary on May 26, 1897, complaining of "fits."

History of Present Illness. Patient appears to have been free from any nervous disorder until a year ago, when, without warning, she fell down, lost consciousness, and remained for twenty minutes in what her friends called a fit. There was no return of "fits" until six months afterwards, when she again had an "attack" described by her friends as quite similar to the first. After this, the fits occurred pretty frequently, lasted from fifteen to twenty minutes, and in all of which she was totally unconscious. Three months ago, after one of these "attacks," she remained unconscious for five days, and during which time there occurred twenty fits (as informed by her friends), while, on regaining consciousness, she found there was considerable weakness of the right arm and leg.

She remained then free from the fits for six weeks, when a fit of a different kind developed, which recurred almost every

ten minutes, and which, she says, corresponds almost entirely with those occurring at present. With these latter fits there is no loss of consciousness, but while it lasts, and indeed for some time afterwards, she loses the power of speech. Occasionally before the commencement of the fit she has a strange sensation in the throat. She notices that the head is drawn over to the right side, and that the arm and leg of the right side are thrown into violent jerking movements. They last only for one to two minutes. Apart from these fits she feels very well. The paresis which resulted some three months ago soon disappeared, and she can now walk perfectly and can use the right arm and hand as freely, she thinks, as the left.

She has never complained of ophthalmic headache, nor of any painful spot on any part of the cranium. Her digestion is good, and the bowels act regularly. She feels that she would sleep well if it were not for the "fits." Her eyesight is perfectly good. She has no abdominal pain, and no swelling of any part of the body. The urine is passed regularly, and she has not considered it unusual either in respect to its quantity or colour. During the fits she passes neither urine nor fæces into bed.

History of previous health. Patient has been a remarkably healthy woman, and apart from a pretty severe attack of rheumatism complicated with pleurisy, which occurred seven years ago, she does not remember having had a day's illness until the present trouble began.

Family history. There is a distinctly tubercular history manifested, as her father and sister, and apparently her mother also, died of phthisis pulmonalis. She has three brothers and three sisters alive and well. No history of insanity or nervous disorder of any kind.

Habits. She is married, but has had no children nor miscarriages. She denies having had any venereal disease, although she remarked that at one time her hair fell out very freely. There was no other indication, however, of any syphilitic taint.

Present condition. Patient is well nourished, and though somewhat pallid, to all appearances looks in ordinary health. There is no lack of adipose tissue, and the muscles are in no way atrophied or flabby. There is, however, a want of intellectual brightness in the eyes. The expression is dull and torpid, and she is slow in answering questions. There is no mental defect.

The pupils are equal, circular, slightly dilated, but respond correctly to light and in accommodation. The tongue is dry,

and deeply fissured transversely—it is protruded in the middle line. Temperature is 97·8° Fah. Pulse is soft, easily compressible, and numbering 114 in the minute. There is no œdema of any part.

Heart is normal as regards percussion and ausculation.

Lungs are practically normal, although a few fine râles are heard at the bases, which accounts for a temporary cough and expectoration which at present exist. Nothing of note is discovered in the *Abdomen*. No movable or enlarged kidney; splenic and hepatic areas of dulness are within normal limits.

Urine is of average quantity, of a dark amber colour; acid reaction; faint mucous deposit; specific gravity 1022; no trace of either albumen, sugar, or blood; microscopical examination discovered a few mucous cells.

While the above report was being written, patient was seized with unilateral convulsions, which occurred almost every eight to ten minutes. They were all almost identical in their nature, they lasted from one and a half to two and a half minutes; commenced almost invariably with a twitching of the small muscles on the right side of the face (not entirely limited to this side however); there was then conjugate deviation of the eyes and face to the right side, shortly afterwards, but in some cases almost simultaneously with this deviation, a complicated movement of the right hand and arm commenced. All the fingers, except the first, are flexed at the metacarpo-phalangeal joints, while the terminal phalanges are in extension. The index finger thus assumes the pointing position. There is then a slight twisting movement at the wrist, the forearm is next slowly pronated and slightly flexed on the arm. During this period, patient frequently mutters to herself. The right leg becomes likewise slightly flexed at the knee joint.

This portion of the attack takes from twenty to thirty seconds when the clonic stage commences, *i.e.*, convulsions proper. There are twitchings of the muscles of the face to a slight extent, chiefly those of the mouth, but almost immediately there are clonic spasms of the muscles of the fingers and thumb of the right hand, then of the whole forearm and arm, so that the right upper limb is thrown into a series of irregular jerking movements. A few seconds after the onset of the convulsive seizure in the arm, the right leg gets convulsed, and is also thrown about in an irregular fashion. At first the convulsion affects the right side alone, but soon after the left arm is slightly affected.

The convulsive seizure commences quietly, but the movements gradually become more intense, reach a maximum, and then gradually subside.

The left arm, when affected in the attack, ceases to be convulsed first, then the right leg, then the arm, and finally the head and eyes rotate backwards into their normal position.

This description, I think, bears out pretty accurately what occurred, although in some fits there were slight modifications. For example, the movements of the arm seemed in some instances to precede the twitching of the muscles of the face, and conjugate deviation of the face and eyes to the right side. Still, I feel sure that in the majority of the fits, the face was first affected, then the arm, and finally the leg, although I have said that in some the arm appeared to be first affected, then the face, and lastly the leg.

The left arm, on occasions, did take part in the movements, but still, these were not of a convulsive nature, at least they did not correspond at all to those of the right.

The right arm was the part which, undoubtedly, took the greatest share in the convulsive seizure. She was perfectly conscious during the attack, but she could not speak. On occasions I asked her questions during the attack and she frequently said "yes," but this was the only word that could be got from her.

Two minutes after the cessation of the convulsive seizure she was able to talk quite distinctly and fluently, although slowly. During this attack the pupils dilated somewhat; the conjunctival reflex was never abolished; respiration and pulse rate increased in frequency; urine and fæces were not involuntarily evacuated.

She seemed to have no warning, other than a "gurgling sensation" in the throat. There was no scream, and no biting of the tongue or foaming at the mouth.

I could discover no paresis of the muscles of the face, arm, hand, leg or foot. Sensation was normal. Knee-jerks present, but not exaggerated. No pain on pressure of muscles, or along course of nerve trunks. No pain on palpation or percussion of the head.

The circumstances of interest were noted in the Journal from time to time, and I shall summarise them here.

May 27, 1897 (day after admission).—To-day, the day after her admission into hospital, patient informed me that she could not move her right arm or leg. She discovered this herself acci-

dentally after one of the fits. The nurses said that the fits had just been of the usual kind, and had occurred every ten minutes on an average. I know that she never lost consciousness, but I am entirely unaware at what time the paralysis set in, and whether it was gradual or sudden. *The arm and leg lay helplessly by the side.* There was no loss of sensation, but she failed to distinguish the difference between the point of the finger and point of a pin on the right side. Apart from this paralytic condition of the right arm and leg, and to a slight extent of the right lower muscles of the face, she had no other unusual symptom. Speech was only affected during and immediately after the convulsions.

June 2 (seven days after admission).—Fits still persist almost as frequently as above noted. She can, however, now move the right foot slightly, although she fails at any attempt to move the right arm or hand.

June 9 (fourteen days after admission).—Since last note fits have gradually become less frequent. She has been taking large doses of bromide of potassium and the syrup of chloral hydrate.

Reactions to faradism, both as regards sensation and muscular contraction, are similar on the two sides.

The right leg can be moved with ease; the right arm not at all.

June 10 (fifteen days after admission).—A recurrence of fits of a milder degree than those noted before, when the face and arm of the right side alone shared in the convulsive storm.

The right leg and left side of the body were in no way affected. The right hand and arm were undoubtedly the centres of the convulsive storm in each case, whereas the twitching of the muscles of the face and conjugate deviation of the head and eyes to the right side were less constantly present. The movements of the right arm and fingers of the right hand correspond entirely with the description given above.

The right arm and hand are completely paralysed; the right leg and foot can be readily moved. No anæsthesia, but *distinct analgesia of the right side of the body.*

June 12 (seventeen days after admission).—Since the 10th patient has had large doses of KI., KBr., and Am. Br., with the result that these milder fits above mentioned soon disappeared. She has had no fit since the night of the 10th.

The right leg has regained so much power that she to-day endeavoured to walk a little, with assistance, in the ward. She can now move the fingers of the right hand, and elevate the arm

at the shoulder. *No albumen in the urine.* Patient feels very well.

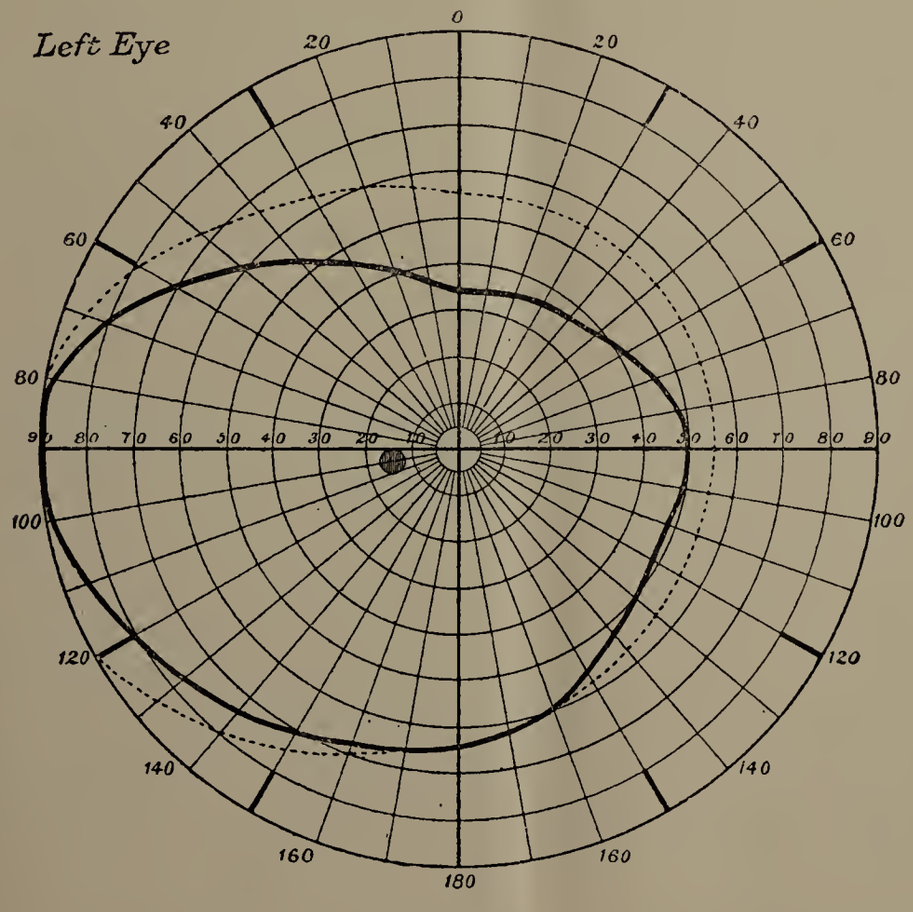
June 24 (twenty-nine days after admission).—Improvement has been steadily progressive. She walks in the ward every day, and the power of the right hand and arm is rapidly returning. No return of fits. Urine normal.

July 1 (thirty-six days after admission).—There is only a slight difference manifest in walking as regards the lower limbs, but it is evident that there is still a slight amount of analgesia of the right limbs. Tactile sensibility is normal.

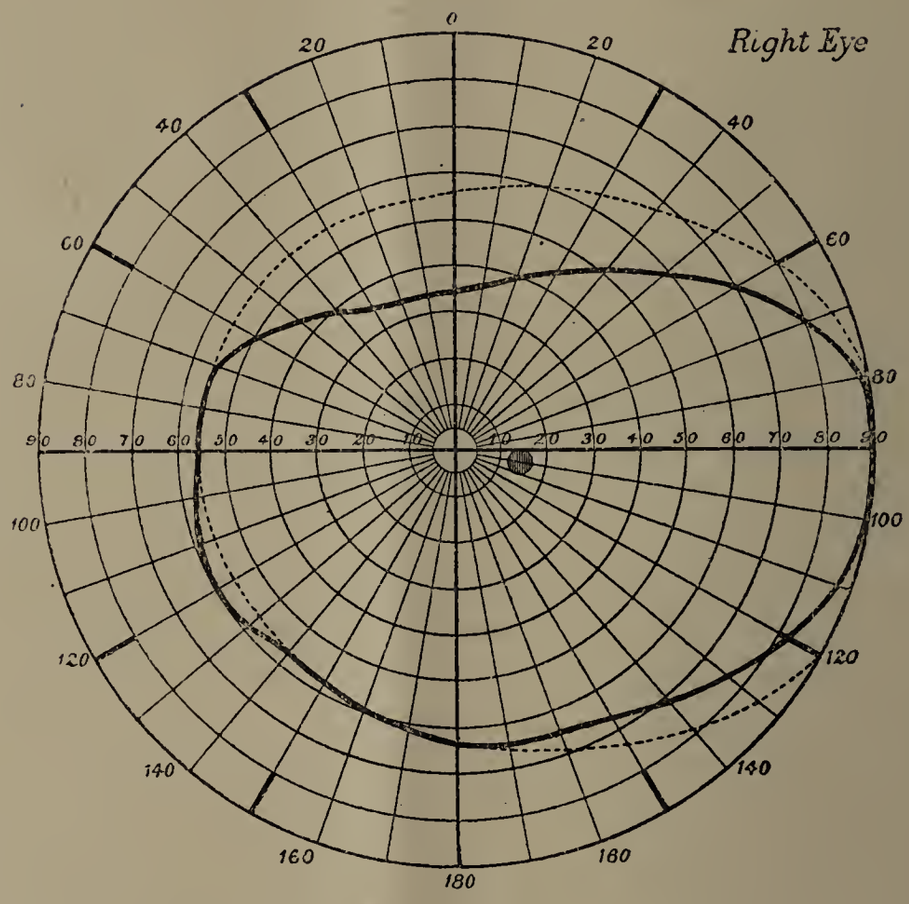
At the tip of the second finger in the two hands the distance by Weber's test is from ·3 to ·5 c.m. *Fundus of both eyes found to be normal.*

July 5 (forty days after admission).—Field of vision was tested to-day by means of the perimeter, and found practically correct on the two sides.

Chart of visual fields of both eyes :—



July 8 (forty-three days after admission).—To-day patient became sick, and she could not answer questions. On examination it was found that she failed to move the *left* arm. The right arm, which was originally paralysed, she moved quite readily.



July 9 (forty-four days after admission).—All signs of paresis of left arm gone.

July 10 (forty-five days after admission).—To-day there was a recurrence of the old fits—of exactly similar nature, but which lasted longer.

July 14 (forty-nine days after admission).—Patient has been menstruating for the last four days, and during which time she has had from eight to nine fits. On the 12th and 13th the fits resembled more the "Idiopathic" epileptic nature.

There was conjugate deviation of head and eyes to the right side, followed first by unilateral convulsions of the right side, which, however, soon after became general. The breathing was stertorous; the face livid; there was foaming at the mouth, and slight bleeding from the tongue and gums. She was quite well in from two to three minutes after coming out of the fit, and she had no drowsy stage, and no loss of control over the bladder and rectum.

July 23 (fifty-eight days after admission).—Patient has had no fits for over a week. Paralysis has almost gone, as she can walk perfectly, and perform the act of writing with apparent ease.

She was dismissed to-day well.

August 6.—Patient had been advised to return to hospital

during her next menstrual period, and so presented herself to-day. *Since last dismissal she has been well.*

August 10.—To-day with the right hand she turned the scale of the dynamometer to 35, and with the left to 55. She has had no fits, but she has been kept well under the action of the bromides.

Dismissed to-day well.

December 23, 1898.—*To-day, over sixteen months having elapsed since she was last dismissed from hospital*, patient was brought to the infirmary by her friends in a speechless and semi-unconscious condition.

The following history was given us by her friends :—It appears that from the time of her dismissal until yesterday, December 22, she remained to all appearance in perfect health, free from fits, and able to attend to her duties.

During this time she suffered neither from headaches, vomiting, pain after food, palpitation, breathlessness, dropsy of any part, pain in the abdomen, disorder of the bowels, retention or suppression of urine, or frequency of micturition ; in fact, her friends state emphatically that she complained of nothing. Her appetite was good, and, indeed, as reported by Dr. Hutton, " might have been justly characterised as ravenous, particularly for nitrogenous food."

Although she appeared somewhat stupid on December 22, she took her dinner and supper heartily, and went to bed. She had had no shiver, or anything to have suggested the onset of an acute illness. At 2 a.m. on the morning of December 23 she was found to be in a fit, livid, and with " jerking movements of the right arm and leg, and snapping of the jaw." These convulsive seizures occurred very frequently, but her friends cannot say definitely how many she had. They were similar to those that occurred sixteen months ago. She was brought up to hospital at 6 p.m. on December 23.

A careful description of the fits was made at the bedside, and the following summary may be said to include all that occurred :—The convulsions were unilateral, confined to the right side, although in one fit the muscles of the face and arm of the left side appeared to participate in the spasm. The eyes first rotated to the right side, with nystagmus-like movements of the eyeballs ; then the head rotated to the same side, with slight clonic movements, more especially of the levator labii superiors and zygomatics of the same side. The right arm was next affected, apparently in an almost identical manner to that fully

described above, and lastly the right leg. Between the spasms she is apparently conscious, as she endeavours with the left hand to cover herself with the blankets. She cannot speak. The pupils are contracted, but equal. She is rather pale, but there is no puffiness of the face, although there is slight pitting on pressure over the tibiæ. No glandular enlargement.

Temperature is 101·4°, pulse soft, rapid, and numbering 142 per minute. Respirations are 32 in the minute. There is occasionally a period of respiration somewhat akin to the Cheyne-Stokes type. *There is a distinct paralysis of the muscles of the face on the right side, and complete paralysis of the muscles of the right arm and leg.* Plantar reflex is absent on the right side; knee-jerks are absent, and there is no ankle clonus.

There is complete anæsthesia of the right side of the body, and partial anæsthesia of the left arm. Heart and abdomen normal. Apart from a few rales at the extreme bases of the lungs (which were noted on last admission), these organs are likewise normal.

All the objective and subjective symptoms of the above report correspond pretty well with the report of her previous admission, but we have still to examine the urine, where we find proof of an acute affection of the kidneys. The first specimen examined contained albumen to the extent of ·1 per cent., urea 6 grs. to 1 oz. blood, a sediment which showed the presence of blood cells, epithelial tube casts, and uric acid crystals. The second specimen drawn off by catheter on the following morning, December 24, the day of her death, showed albumen to extent of ·9 per cent., a large quantity of blood, abundant epithelial, and other tube casts.

Hypodermic injections of pilocarpine were tried with only temporary benefit, and patient gradually passed into a condition resembling the "status epilepticus," and died at 3.50 p.m., December 24, 1898, after a two days' illness.

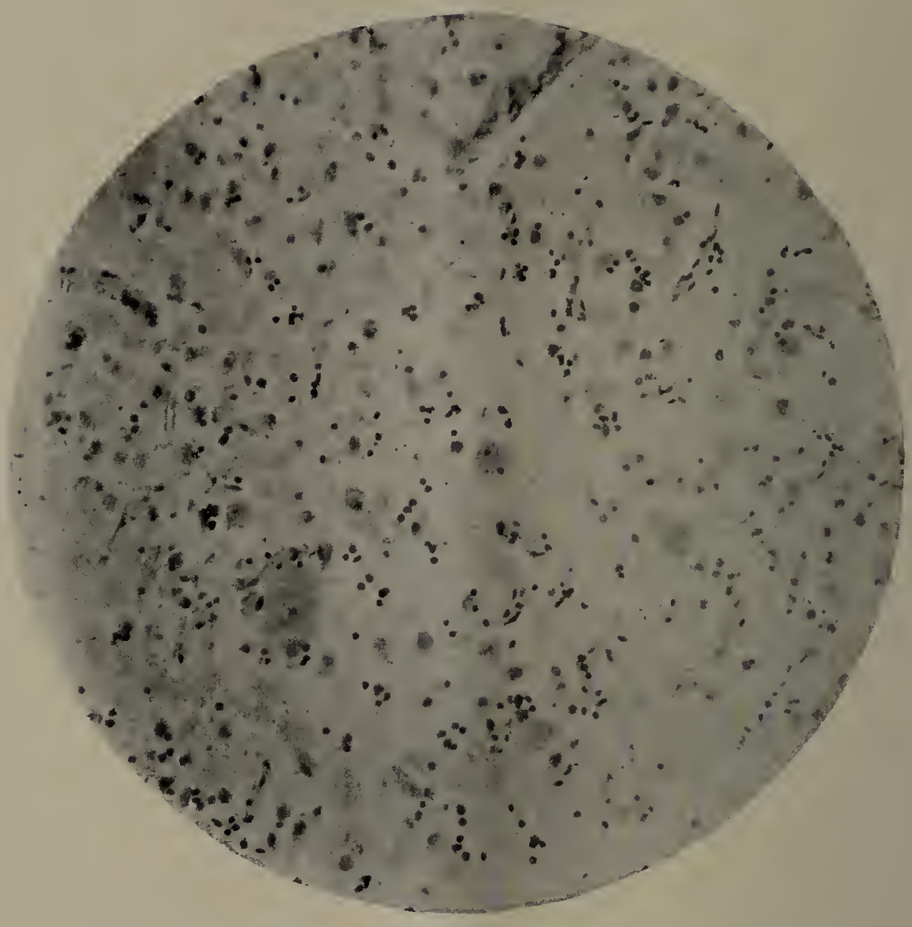
The temperature gradually rose after her admission, and shortly before her death it was recorded as 104° F.

A post-mortem examination was made, the results of which are briefly as follows:—

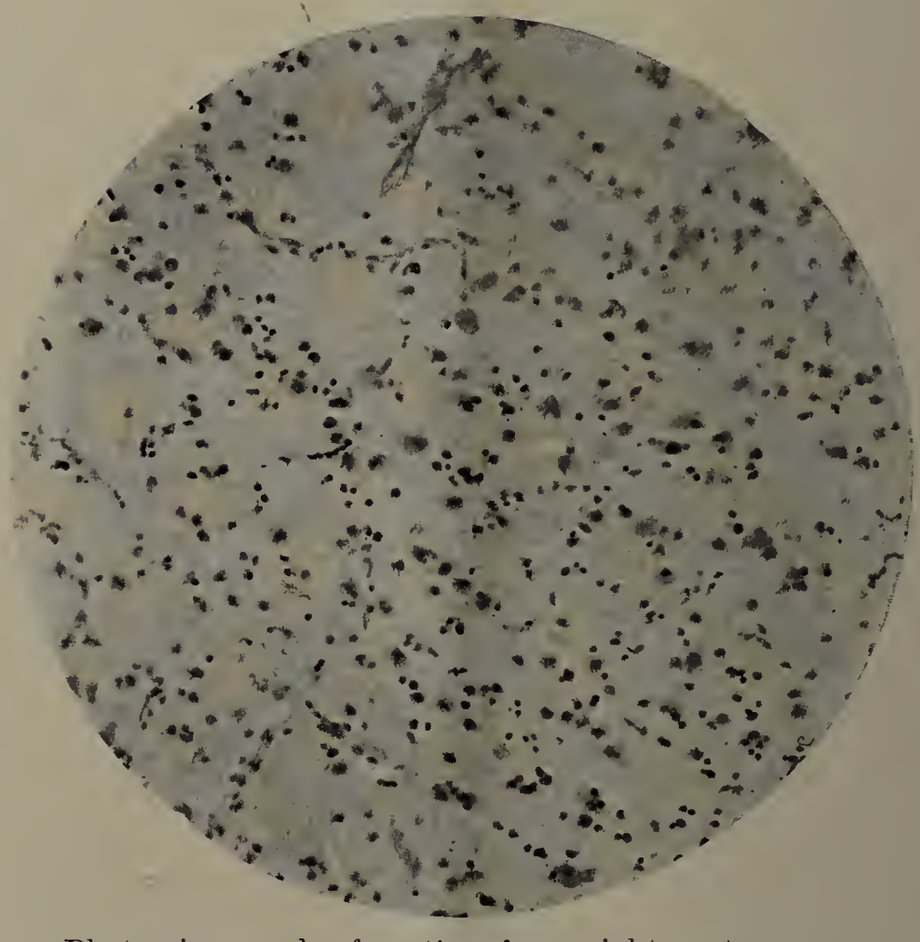
The heart is somewhat fatty, but there is no insufficiency of the valves.

The lungs are non-adherent, but considerably œdematous. Spleen and liver are large and soft with *post-mortem* staining. Stomach, intestines and pancreas are normal.

The brain is superficially congested posteriorly (*post mortem*). *Its substance shows nothing abnormal.* Dr. Dittmar in his report



Photomicrograph of section from left motor area.



Photomicrograph of section from right motor area.

says: "There is nothing unusual in the appearance of the microscopical sections which I have prepared from both motor areas of the brain—if anything there appears to be a preponderance of capillaries in that of the *right* side."

The kidneys present appearances of an acute cortical nephritis, accompanied by profuse desquamation and destruction of epithelium. The adrenals, ureters and bladder are normal.

I have described this case as fully as possible, in order that the various points of interest and difficulty with regard to its probable pathology and diagnosis may be criticised, and I shall now shortly discuss some of the problems that presented themselves under the headings of symptoms, diagnosis, ætiology and pathology.

Symptoms.

Each fit was almost invariably ushered in by (1) *conjugate deviation of the eyes and head* to the right side. The axes of the eyeballs remained absolutely parallel. The movement was gradual, and free from any spasmodic jerking; moreover, there was definite precision in the commencement of rotation of both eyes and head. We know from the writings of Broadbent (33), Ross (34), and Gowers (35), how this symptom depends on some change in the opposite side of the brain, frequently produced by a hæmorrhage. If the muscles of the side opposite to the lesion are paralysed, then the antagonistic muscles draw both eyes and head to the non-paralysed side, *i.e.*, the head turns, and the eyes look towards the side of the lesion. However, if there is an irritation of one motor area, or rather of that portion of the cerebral cortex that has to do with the lateral deviation of the eyes and head, then the muscles of the side opposite to the lesion contract, and cause rotation of the eyes and head to the paralysed side, *i.e.*, the head turns away, and the eyes look from the lesion. In this case, there was probably an irritation of that area which lies immediately in front of the truly motor zone (see diagram of Beevor and Horsley); and, as a result, the nucleus of the sixth nerve of the right side was irritated,

which produced contraction of the right external rectus, directly and indirectly, through the communicating fibres from this nucleus to the third nerve and spinal accessory of the opposite side, produced contraction of the left internal rectus, and left sterno-mastoid, although the right inferior oblique muscle (as described by Ross) also probably contracted. I consider that this area was the first to become irritated, and, owing to its physiological situation, this fact accounted for the convulsive seizures that depended on irritation of the neighbouring areas for the face and upper extremity. There was never any sign of paralysis of the orbital muscles, as is frequently the case from certain brain tumours.

(2) *Hemiplegia* occurred, not as is usually described by Hughlings Jackson, but in an irregular fashion, and with apparent suddenness. As we have above noted, hemiplegia after unilateral convulsions depends upon the weakened state of certain motor nerve cells after the explosion, which produced the convulsions, had taken place. One would expect to have a gradual hemiplegia resulting from a gradual exhaustibility of the motor cells due to frequent explosions of nerve energy. In this case we must consider the form of epilepsy as one of the apoplectiform or congestive type. The history obtained from patient's friends tells us of the suddenness of the onset of fits, the period of over a week's unconsciousness, the resulting hemiplegia of the right side of the body—on her admission we observed the occurrence of numerous fits with no evidences of hemiplegia, but, on the day following, the development of paralysis of the right arm and leg—and, again, on her last admission, we noted hemiplegia with paralysis of the lower muscles of the face on the right side.

It is not likely that in this case there were actual hæmorrhages in the cerebral cortex, as there would have been evidences *post mortem*. It is quite probable that after a more severe convulsive attack, the vessels in the neighbourhood of the left motor area became contracted, and a congestion resulted, allowing of a greater disturbance to the nutrition of these motor nerve cells.

It is difficult to account for the very fleeting paralysis of the left arm that occurred on one occasion, as this was preceded by no convulsive attack.

(3) *Aphasia* occurred shortly after the commencement of each fit, and lasted, as a rule, for two minutes after the cessation of the convulsive seizure. Aphasia, as has been graphically described by Hughlings Jackson, occurs almost invariably when the spasm begins in the right side of the body. In this case, the defect of speech was not due to difficulty of articulation, depending on paralysis of the labial muscles, but was from a true aphasia, the result of some change in the speech-producing centre on the left side of the brain.

Todd, in his lectures on Diseases of the Nervous System (lecture xv.), says:—"There is a peculiar class of cases of epileptic hemiplegia, in which the exciting cause of the epileptic fit at the same time damages or greatly injures voluntary power and speech." A factor in support of the view which I have expressed above, that the convulsive storm appeared almost invariably to start in the face, is the fact that Hughlings Jackson observed that aphasia frequently occurred when the face was first affected, but rarely when the convulsion commenced in the arm or leg.

An interesting case is reported by Sorel (36), where the symptoms were in many respects similar to the one at present under consideration. In Sorel's case the convulsions started in the face, but were preceded by conjugate deviation of the eyes and head. There was also *aphasia.* This transitory aphasia has been noted also by Audrey, Weber, Brown Sequard, and others.

(4) *Loss of consciousness* is rarely present in true Jacksonian epilepsy, and the victim of these convulsive seizures can, as a rule, describe accurately the general march of the storm, and repeat the various events that happened during the attack. Apart from her severe illness at the commencement of the disease, when her friends described her as unconscious for a week, and apart from her condition towards the end of her illness, which resembled the *status epilepticus,* she was perfectly conscious during all the convulsive seizures.

(5) *Disturbances of sensation* are rare accompaniments of Jacksonian epilepsy, whether there is post-epileptoid paralysis or not. I think one of the most important and interesting features of this case was the presence of analgesia of the right leg, and less markedly of the right arm, during most of her illness, and the complete anæsthesia of the whole right side of the arm and leg during the latter days of patient's life.

We know that anæsthesia does occur in some forms of Jacksonian epilepsy, depending on a syphilitic affection, but in these cases it is disseminated, occurring in patches over the backs of the hands, and perhaps outer aspects of the leg (Fournier, Barbier). But, in this case, we had no definite history of syphilis, and certainly *post mortem* there were no changes at all suggesting such a condition.

The question of anæsthesia and analgesia in relation to cerebral localisation has been put to experimental test by Ferrier, Schäfer, Horsley, and Yeo. All experiments show that destruction of the motor areas of the brain, however extensive, does *not* produce either anæsthesia or analgesia. These observers have found that the cerebral centres for such sensations lie in the hippocampal and inferior temporal regions, and also in the gyrus fornicatus.

I consider that in this case, the presence of analgesia, and anæsthesia shortly before death, depended most probably on certain changes that had taken place in the communicating fibres that connected the tactile centres with those of the motor region. Few cases are recorded where anæsthesia was present with unilateral convulsions, but one described by Broadbent (37) is of great interest. He records a case of epileptoid convulsions with hemi-anæsthesia, and in discussing the case, he says:—"Another very interesting feature in the case was the loss of sensation, which for a time was absolute in the right hand and arm, and well marked, though not complete in the body and leg of the same side." As this patient was cured under large doses of iodide of potassium, it is possible that this case belonged to the syphilitic group, where convulsions and anæsthesia have been frequently recorded.

Diagnosis.

Whenever convulsions are unilateral, occurring seldom, and it may be in rapid succession, where definite parts of the body are affected, and where the convulsive storm is ushered in by the contraction of a particular group of muscles, with no loss of consciousness, at any rate at the commencement of the spasm, then we are dealing with a case of Jacksonian epilepsy. It matters little what the cause may be. The essential feature in such cases is the fact that a particular motor area, or only a portion of it, is irritated. So that, if in hysteria, when we speak of a functional state, definite unilateral convulsions exist, the case is one of Jacksonian epilepsy, provided that the seizures are not feigned.

Again, if in uræmia the convulsions are unilateral and consequently depend on irritation of the motor area by urinary elements, the same diagnosis may be arrived at. The two sets of cases might be talked of as hysterical and uræmic convulsions, assuming the Jacksonian type.

Still, other symptoms being taken into account, we are able to distinguish the disease from others, more especially from hysteria, true epilepsy, hystero-epilepsy, and uræmia.

(*a*) *Hysteria*.—In very few respects did our patient's condition present the characters of the "hysterical attack."

There was no emotional utterance before the seizure—no sudden falling to the ground with feigned loss of consciousness—no subsequent coma and fits of laughing and crying. There was no drooping of the eyelids. There was, however, hemiplegia and sensory disturbance, affections which are frequently present in hysterical patients, but in such cases one never meets with convulsions which pursue an almost exact course in each seizure, as in the present case.

(*b*) *True epilepsy*.—Although at one time the fits more closely resembled the truly epileptic nature, as a rule they largely differed from them. There was no history of epilepsy or other neurotic disease in the family—patient had had no convulsions or fits as a child—unconsciousness

practically never existed—there was rarely biting of the tongue, and at no time was there loss of control over the bladder and rectum. Moreover, the fits were localised, and were almost invariably unilateral. On one or two occasions, due to the severity of the storm, the left arm participated in the attact, but the fits may be definitely said to have never been general. In four minutes or so after the seizure she was well.

(*c*) *Hystero-epilepsy.*—Charcot (38) in one of his lectures describes this disease as one of a truly hysterical nature, but which assumes, owing to the severity of the hysteria, an epileptoid condition. He entirely dismisses the theories that have been suggested of an admixture of the two diseases, hysteria and epilepsy. If this be correct, then we are at a loss as to the cause of the irritation that produces the epileptoid attacks.

In hysterical patients there may be a "*functional neurosis,*" but this functional neurosis implies, on physiological grounds, change (it may be temporary) of nerve elements. However, in hystero-epilepsy, the convulsions correspond to the idiopathic epileptic type, and are not localised as in Jacksonian epilepsy.

Besides, there is, as a rule the hysterical crying at the commencement of, and the cataleptic state after, the convulsion. Although we are not aware of the change that occurs in the brain as a result of hysteria, it is quite within the range of possibility that hysteria, which may assume the truly epileptic nature by disturbance of the whole cerebrum, may as readily assume the Jacksonian type by irritating one motor area. Such a disease might well be called *Hystero-Jacksonian epilepsy.*

By this term, however, I intend to denote hysteria as the primary disease with, as a sequel, irritation of one motor area from some nerve change, the cause of which is as yet unfathomed. At the same time, I fail to see why one motor area could be so isolated.

It is easy to distinguish Jacksonian epilepsy from the hystero-epilepsy of Charcot, but when we endeavour to distinguish it from the disease which I have designated as

hystero-Jacksonian epilepsy, it is more difficult, especially when we consider certain points in the history, and symptoms of our patient, and the absence of any macroscopic or microscopic change in the motor zone.

I do not, however, uphold the idea of hysteria as the essential disease in this case, but whether hysteria be an element in the case or not, it matters little, for the disease is one of Jacksonian epilepsy, whatever be the cause.

(*d*) *Uræmia.*—We are aware that in certain forms of Bright's disease, the first symptoms are ushered in by convulsions. They may be epileptic in character, and indeed well localised and unilateral (39).

Had the disease in the present case depended on a chronic nephritis, no symptoms of which had presented themselves when the convulsions began, there would almost assuredly have been the development of symptoms during the long interval that elapsed between her second and last admission into hospital. Although the convulsions in uræmia at times do resemble Jacksonian epilepsy, and although French writers discuss a toxic epilepsy depending on the action of the urinary elements on one motor area, as a rule the convulsious are less accurately defined, and do not pursue the course that has been described.

The only place that we can give to uræmia is in the latter stages of patient's life, when an acute nephritis developed, these urinary elements acting on the already changed or susceptible motor area of the brain.

Ætiology and Pathology.

Had there been any macroscopical or even microscopical change in the left motor area of the cerebrum, it would have been an easy matter to discuss the ætiology and pathology of this case. The accompanying photomicrographs and report by Dr. Dittmar on the histological appearances negative any such change. For this reason the case is all the more interesting, even though any theories or views that may be advanced are speculative.

That the left motor area of the brain was irritated, and

at certain times excessively so, is a fact that must be accepted.

The symptoms that presented themselves were in all respects in harmony with our knowledge of unilateral convulsions depending on an irritation starting at a definite point in the motor area, and spreading from that region to the parts around in definite order. The irritation started in the area for lateral deviation of the eyes and head, passed across to the centre for the face, interfering with the communicating fibres that had to do with tactile sensibility and the speech-producing centre, then passed up the fissure of Rolando to the centre for the arm, and then for the leg.

As there was no tumour, change in walls of arteries, cicatrisation, or evidence of ramollissement present, we must fall back on one of two theories as the cause of the convulsive seizures, the one vaso-motor, the other chemical. The first of these would account for the apoplectiform nature of certain of the fits, due probably to a contraction of some of the smaller arteries producing a congestion of the surrounding neuroglia; this might cause effusion which disappeared again by absorption, and left no trace *post mortem*.

The other theory, chemical in nature, has been already described in the remarks at the commencement of this paper on the general pathology of Jacksonian epilepsy. Either of these theories is possibly correct, and yet it is difficult to conceive why in both instances there should be vaso-motor and chemical disturbances in so localised an area.

Granted then that from either of these causes, or possibly from some other, the left motor zone, or part of it, is changed in nervous structure, or rendered susceptible, it is easy to understand the remaining difficulties that presented themselves. The convulsions ceased after a time, and were set agoing by the next menstrual period. This then might be an exciting cause which acted reflexly on the previously changed or susceptible area.

We might possibly have a *reflex Jacksonian epilepsy* in this instance. But after over a year's cessation of the

fits, and owing to the occurrence of an acute nephritis and the absorption of urinary elements into the blood, *a toxic Jacksonian epilepsy* occurred, depending on the action of these elements on the old standing susceptible left motor area of the brain.

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